

Acquired Heart Disease in Childhood

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Outline

- **Acute Rheumatic Fever and Rheumatic Heart Disease.**
- **Kawasaki Disease.**

The background of the slide features a pattern of stylized, overlapping leaves in various shades of yellow and orange, creating a warm, autumnal feel.

ACUTE RHEUMATIC **FEVER**

Acute Rheumatic Fever (ARF)

Definition:

ARF is an inflammatory disease that occurs as a delayed nonsuppurative sequel of group A streptococcal upper respiratory infection.

Definition (cont.):

♥ *In its classic form ARF is acute, febrile, and largely self-limited disorder.*

♥ *It involves joints, heart, brain, and skin in varying combinations.*

Etiology:

ARF development requires an antecedent infection with :

♥ *A specific organism:*

group A streptococcus

♥ *At a specific site:*

the upper respiratory tract.

Etiology (cont.):

- ♥ *There is evidence that group A streptococci serotypes vary in their rheumatogenic potential.*
- ♥ *Strains causing clusters or epidemics usually belong to a limited number of serotypes (e.g. 3, 5, 18, 24, and others).*

Pathogenesis:

Various theories were explaining the mechanism by which group A streptococci elicit the connective tissue inflammatory response that constitutes ARF including:

Pathogenesis (cont.):

- ♥ 1. *Toxic effects of streptococcal products initiating tissue injury.*
- ♥ 2. *Inflammation mediated by antigen-antibody complexes, localized to sites of tissue injury.* causing the menfistation
- ♥ 3. *Autoimmune phenomena induced by similarity of certain streptococcal and human tissue antigens.*

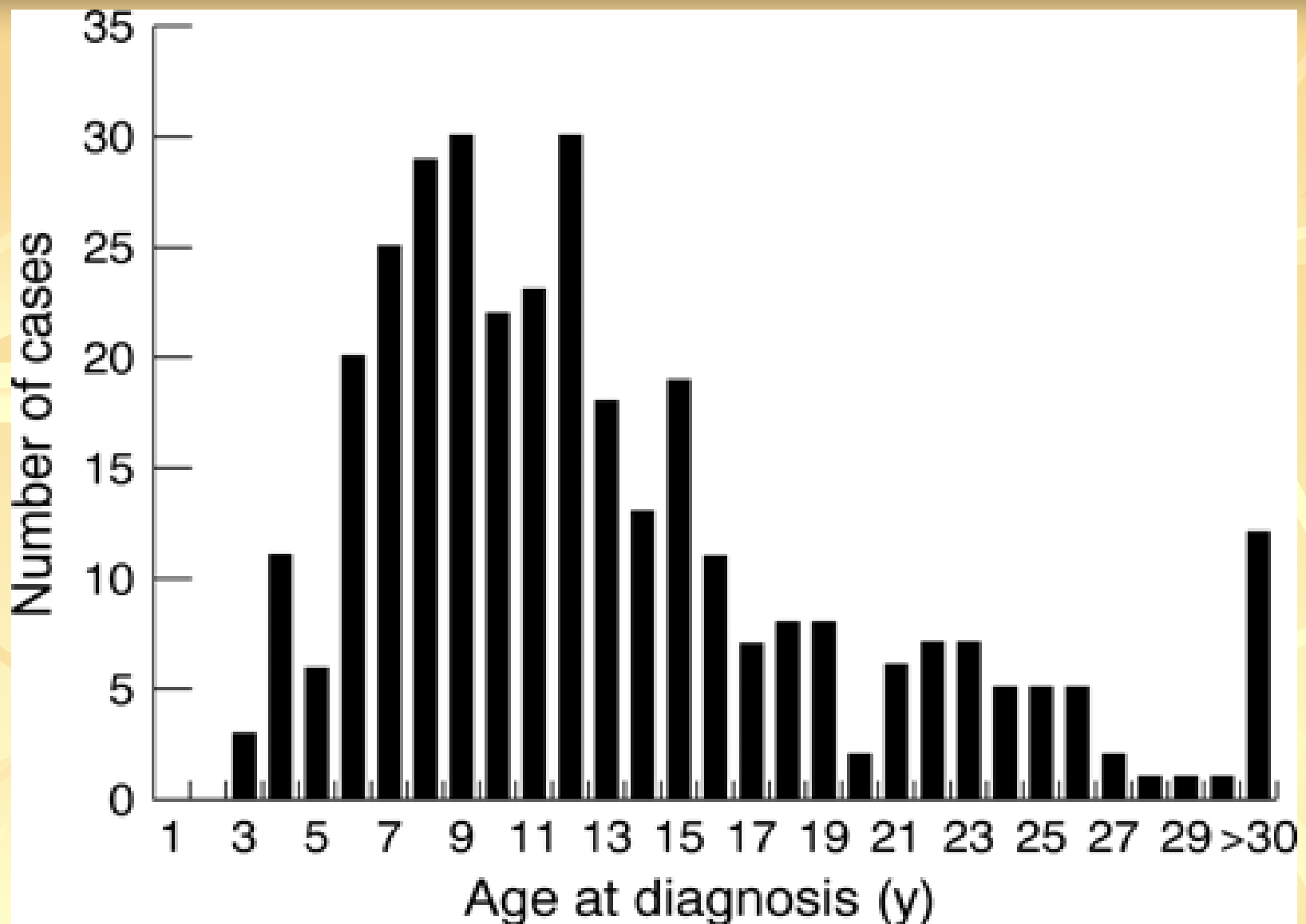
most acceptable skin
rash , endochrsotots
chorea

Pathogenesis (cont.):

Several observations suggest that specific genetic constitution of the host may predispose to the occurrence and / or recurrence of ARF.

ARF Epidemiology:

- ♥ *It mirrors the epidemiology of streptococcal pharyngitis.* can occur in cold
- ♥ *The peak age incidence is 5 to 15 years, but both primary and recurrent cases can occur in adults. ARF is rare before the age of 4 years.* can occur before 5 or after 15 but this is the peak age
- ♥ *There is no clear-cut gender predisposition, although females are more likely to develop certain manifestations such as Sydenham's chorea.*



ARF age at diagnosis in a high incidence population in Australia (Arch Dis Child 2001 ;85:223-227)

ARF Epidemiology (cont.):

- ♥ *There is no known racial predisposition.*
- ♥ *The disease peaks in cooler months of the year.*
- ♥ *Crowding and low socioeconomic status are the main environmental factors favoring the occurrence of ARF.*
- ♥ *ARF remains a common and important health problem in developing countries.*



Crowdedness is an environmental factor favoring occurrence of ARF

ARF Pathology:

Exudative and proliferative inflammatory lesions in the connective tissues of the heart, joints, and subcutaneous tissues, characterize ARF.

Pathology (cont.):



In the heart:

pathognomonic of RF



Aschoff nodules are virtually pathognomonic.

They consist of a central area of fibrinoid surrounded by lymphocytes, plasma cells, and large basophilic cells; some of them are multinucleated.

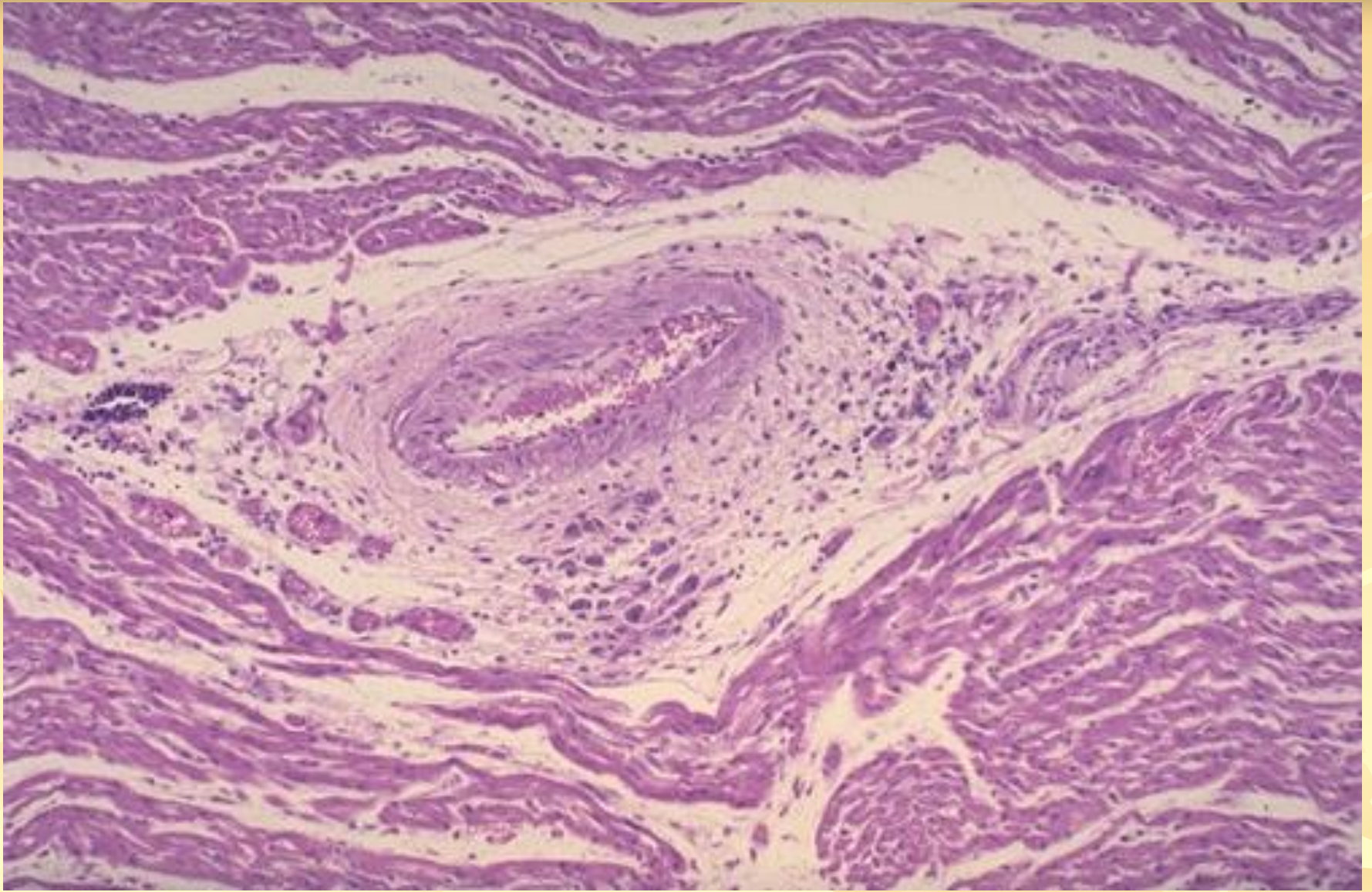


Cardiac findings may involve pericardium, myocardium, or endocardium.

gross pathology



Acute Rheumatic Fever Vegetations



Aschoff Nodule

Pathology (cont.):

♥ *In the joints:*

in bigger joints and no residual arthritis
self limited within 3-4 weeks w/o treatment

♥ *The arthritis of ARF is characterized by a fibrinous exudate and sterile effusion without erosion of joint surfaces or pannus formation.*

ARF is licking the joints

but is biting the heart



ARF Clinical Manifestations:

*The **latent period** between the antecedent streptococcal infection and the onset of ARF symptoms ranges between 1 and 5 weeks (average is 19 days) for both primary and recurrent attacks.*

Cl. Manifestations (cont.):

Five clinical features are so characteristic that they are recognized as major manifestations:

Carditis

Polyarthritits

Chorea

Erythema marginatum

Subcutaneous nodules

Arthritis:

Joint involvement ranges from arthralgia alone to acute disabling arthritis (swelling, warmth, erythema, limitation of motion, and severe tenderness).

painful joint = arthralgia

Arthritis (cont.):

*The **larger joints** of the extremities are usually involved:*

- ♥ *Most frequently: **knees** and **ankles**.*
- ♥ *Frequently: **wrists** and **elbows**.*
- ♥ *Occasionally: **hips** and **small joints of hands and feet**.*
- ♥ *Rarely: **shoulders**, **lumbosacral**, **cervical**, **sternoclavicular**, and **temporomandibular** joints.*

very commonly affected in RA to differentiate



Ankle Arthritis

Arthritis (cont.):

- ♥ *Characteristically, **migratory polyarthritis** pattern is noticed.*
 - ♥ *In most instances, inflammation to any joint begins to **subside spontaneously** within **one week**.*
 - ♥ *The total duration of polyarthritis is **no more than 3 or 4 weeks** leaving **no residual joint damage**.*
- joint pain for 3-4 months ruling out RF

Carditis:

1st endocardium w\ valves

then pericardium ...

- ♥ *ARF may involve endocardium, myocardium, and pericardium (i.e. can induce pancarditis).*
- ♥ *Carditis is the most important manifestation of ARF as it is the only one that can cause significant permanent organ damage and death.*

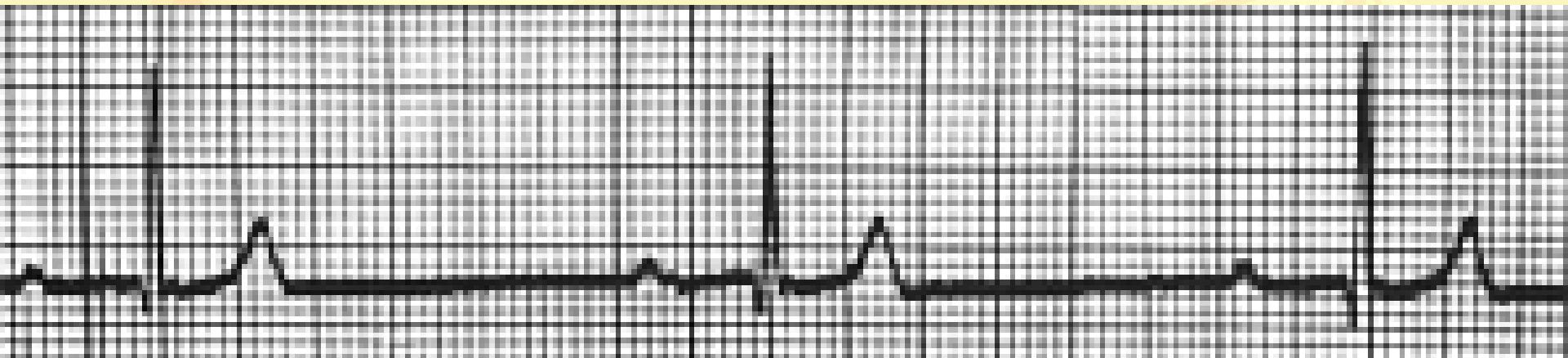
Carditis (cont.):

The diagnosis of carditis requires the presence of one of the following:

- ♥ 1. **New organic cardiac murmur** (e.g. apical systolic murmur of MR, apical mid-diastolic (carey-coombs) murmur of MS, or basal diastolic murmur of AR). most likely carditis
- ♥ 2. **Cardiomegaly.** apex beat and CXR
- ♥ 3. **Pericardial friction rub (i.e. pericarditis) or muffled heart sounds due to PE.**
- ♥ 4. **Congestive heart failure.**

Carditis (cont.):

Different rhythm disturbances may occur during the course of ARF, the commonest of which is first degree atrioventricular block (prolonged P-R interval). normally 3-4 mm ... here 7-8 mm



Sydenham's Chorea:

affection of basal ganglia

It is characterized by:

- ♥ **1. Rapid purposeless involuntary movements in the extremities and face and may sometimes be unilateral.**
- ♥ **2. Hypotonia.**
- ♥ **3. Inability to maintain a tetanic muscle contraction.**

most useful in diagnosis .. ask to extend the arm and only maintain this position for seconds then move involuntarily . even to extend her tongue same thing it's the only manifestation of Rf occur in female more
- ♥ **4. Emotional lability.**

the girls laughing and crying w/o reason

Sydenham's Chorea (cont.):



Chorea usually occurs after a latent period that is longer than that associated with other manifestations of ARF.



Chorea frequently occurs in “pure” form.

only with involuntary movement . if took good hx
mom says: last month couldn't move (bcs of arthrits)
or got skin rash though allergy (that was erthema maginitum)

Subcutaneous Nodules:

♥ *These are firm painless subcutaneous lesions.*

♥ *Varying in size from few millimeters to 2 cm.*

♥ *They occur in crops over bony prominences and tendons.*

groups

behind the elbow or the lower

back around sacrum

u should look for them

in hidden places pt won't

complain

Subcutaneous Nodules (cont.):

- ♥ *They do not appear until at least 3 weeks after onset of ARF and last for 1 to 2 weeks.*
- ♥ *Nodules almost always appear in association with severe carditis.*



Subcutaneous Nodule

Erythema Marginatum:

♥ *The rash begins as an erythematous ^{flat} macule or ^{raised} papule, which extends outward while the skin in the center returns to normal.*

♥ *The lesions may be raised or flat, are neither pruritic nor indurated.*

♥ *They blanch on pressure.*

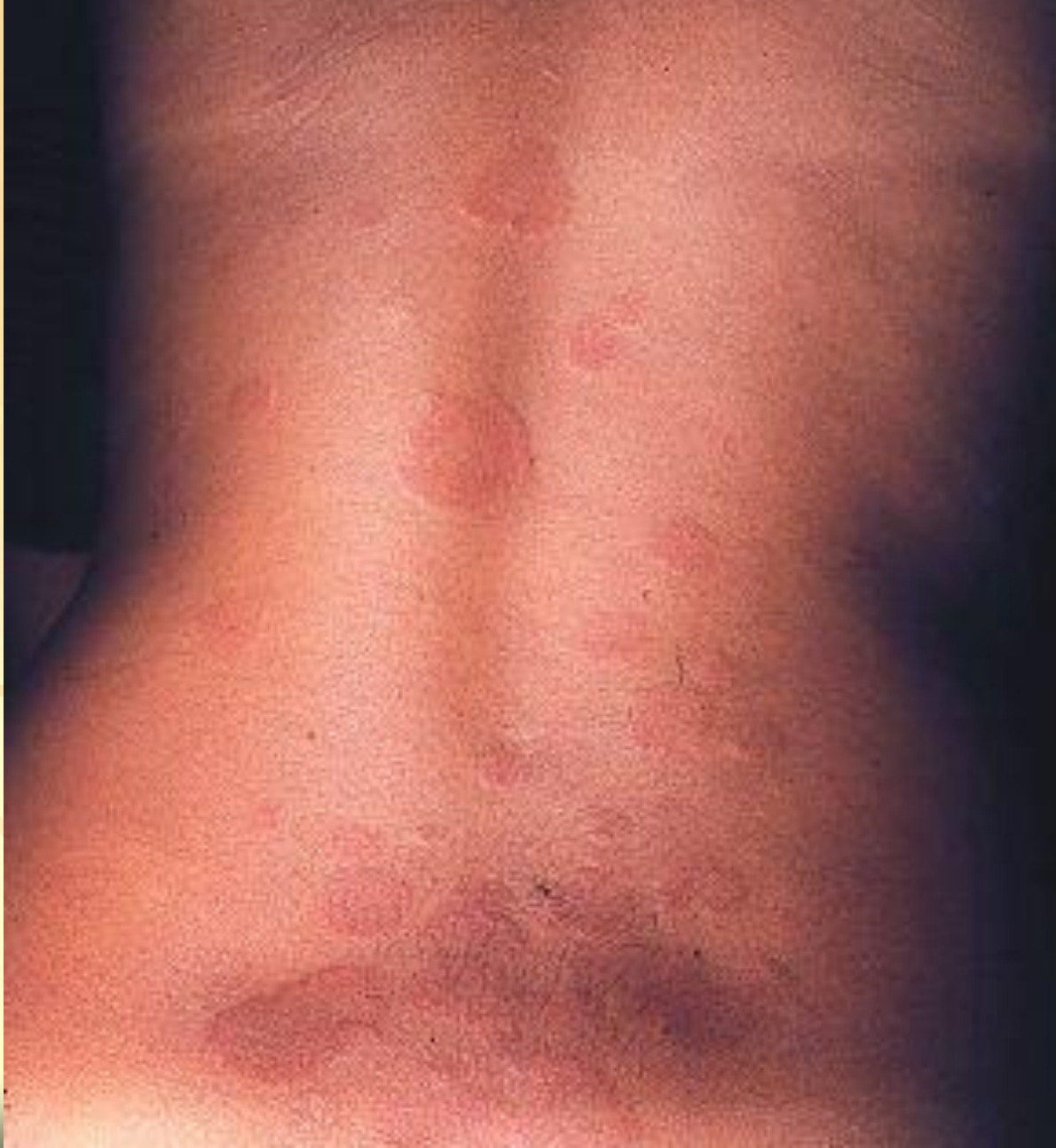
Erythema Marginatum (cont.):

- ♥ *The lesions vary in size.*
- ♥ *They appear in the trunk and proximal extremities, sparing the face.****
- ♥ *The lesions migrate from place to place leaving no residual scarring.*



Erythema Marginatum

Erythema Marginatum



periphery is darker than center

Erythema Marginatum (magnified)

Investigations:

- ♥ *No specific laboratory test is diagnostic of ARF.*
- ♥ *Usually there is polymorph nuclear **leucocytosis** and mild to moderate normocytic normochromic **anemia**.*
- ♥ *Evidence of acute inflammation is prominent including **raised CRP** and **ESR**. not specific but always high with ARF*

Investigations (cont.):

Lab. Evidence of recent group A streptococcal infection are usually there including: mandatory

♥ *Positive throat swab cultures.* will be -ve in RF cus latent perios

♥ *Positive rapid streptococcal antigen test.*

♥ *Elevated or rising antistreptococcal antibody titers e.g.:*

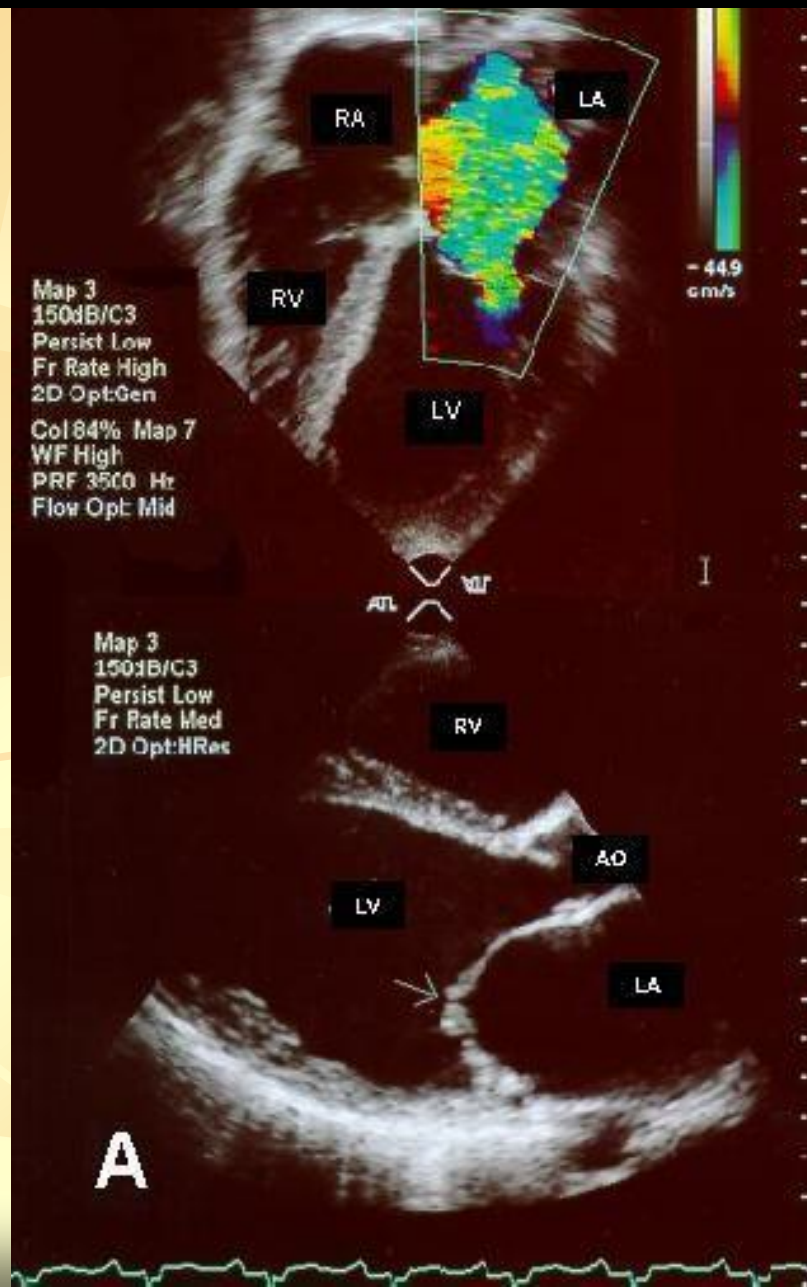
Antistreptolysin O (ASO), ag hard to find but ab will stay for months

*Antideoxyribonuclease B (anti-DNase B),
or Antihyaluronidase titers.*

Investigations (cont.):

- ♥ *ECG may reveal evidence of rhythm disturbance (e.g. prolonged P-R interval).*
- ♥ *CXR may show findings suggesting congestive heart failure or pericarditis.*
- ♥ *Echocardiography may document myocardial and valvular dysfunctions and pericardial effusion.*

Rheumatic Mitral Valve Regurge



Diagnosis:

Modified Jones Criteria:

Two major criteria or one major and two minor criteria indicate a high probability of ARF, provided that there is supporting evidence of recent streptococcal infection.

?? Exceptions ??

rhumatic chorea w\ no need to others

Modified Jones Criteria

<u>Major criteria</u>	<u>Minor criteria</u>	<u>Evidence of recent strept. infection</u>
Carditis	Fever	Positive throat cultures
Polyarthrititis if u have this don't count arthralgia	Arthralgia	Positive rapid streptococcal antigen test
Chorea	Prolonged P-R interval	Elevated antistreptococcal antibodies e.g. ASO , anti DNase B, or antihyaluronidase titers
Erythema marginatum	Elevated acute phase reactants e.g. ESR and CRP	
Subcutaneous nodules		

Differential Diagnosis

Juvenile rheumatoid arthritis

Septic arthritis affecting one joint

Sickle-cell arthropathy

Kawasaki disease

Myocarditis can be confused with pericarditis in RF

Scarlet fever acute tonsillitis not 3 weeks ago

Leukemia

ARF Treatment:

- ♥ 1. *Bed rest for inflamed joints and / or congestive heart failure (CHF).*
- ♥ 2. *Specific therapy for CHF e.g. diuretics.*
- ♥ 3. *Oral penicillin V 40 mg /kg /day for 10 days to eradicate throat streptococci.*

Oral cephalosporin or erythromycin may be an alternative in case of penicillin allergy.

Treatment (cont.):

for the inflammatory process
aspirin in high doses

4. **Anti-inflammatory agents: Aspirin**
*in a dose of 90-100 mg / kg / day in
4 divided doses after food is very
effective decreasing fever, toxicity,
and joint inflammation. After 2
weeks aspirin dose can be reduced
to 60-70 mg / kg / day for an
additional 6 weeks.*

Treatment (cont.):

Corticosteroids are reserved for:

- ♥ *Patients with severe carditis manifested by CHF,*
- ♥ *Patients who are unable to tolerate large doses of aspirin, or*
- ♥ *Patients whose signs and symptoms are not adequately suppressed by aspirin.*

Treatment (cont.):

- ♥ *Prednisolone 2 mg / kg / day in divided doses may be initiated.*
 - very rapid effect
 - pt tolerate more than aspirin
 - incidence of recurrence with RF after corticosteroid much higher than aspirin
- ♥ *After 2-3 weeks it should be withdrawn slowly over an additional 3 weeks period.*
- ♥ *Aspirin is better to be given for one month after discontinuation of steroids to avoid rebounds, which frequently occur after steroid therapy.*

Treatment (cont.):

Patients with significant Sydenham's chorea need specific treatment.

give medical ttt

♥ ***Diazepam** in acute severe cases.*

♥ ***Haloperidol** was frequently used.*

♥ ***Sodium valproate** was found effective and has less side effects.*

ARF Prophylaxis:

1. Primary prophylaxis of ARF:

It consists of accurate diagnosis and appropriate treatment of streptococcal throat infections.

ARF Prophylaxis:

2. Secondary prophylaxis:

♥ It aims at preventing the recurrence of ARF in patients who have already suffered a rheumatic attack.
cardiac valves can't tolerate 2nd attack of RF > will have severe cardiac valve disease

♥ Benzathine penicillin intramuscular injections every 3-4 weeks is recommended.
long acting penicillin, IM,
every 4 weeks if no cardiac for 5 years or till age 21

♥ The dose is 600,000 units in patients weighing less than 27 kg. and 1,200,000 units in patients weighing more than 27 kg.
every 3 weeks for cardiac lesion for life

ARF Prophylaxis:

- ♥ *This secondary prophylaxis should be maintained **indefinitely** for those with rheumatic heart disease.*
- ♥ *Other rheumatic subjects should be protected until they reach age 21 and 5 years have elapsed since the last rheumatic attack, whichever is longer.*
- ♥ *A skin sensitivity test **should** be done before each injection.*

ARF Prophylaxis:

3. Tertiary prophylaxis:

- ♥ It means bacterial endocarditis prophylaxis for patients with rheumatic heart disease whenever they undergo dental or surgical procedures likely to evoke bacteremia.
- ♥ This is not necessary in rheumatic patients free of residual heart disease.
1 does of amoxicilin 1 hrs before procedure
- ♥ Receiving rheumatic fever prophylaxis does not exempt the patient from endocarditis prophylaxis.

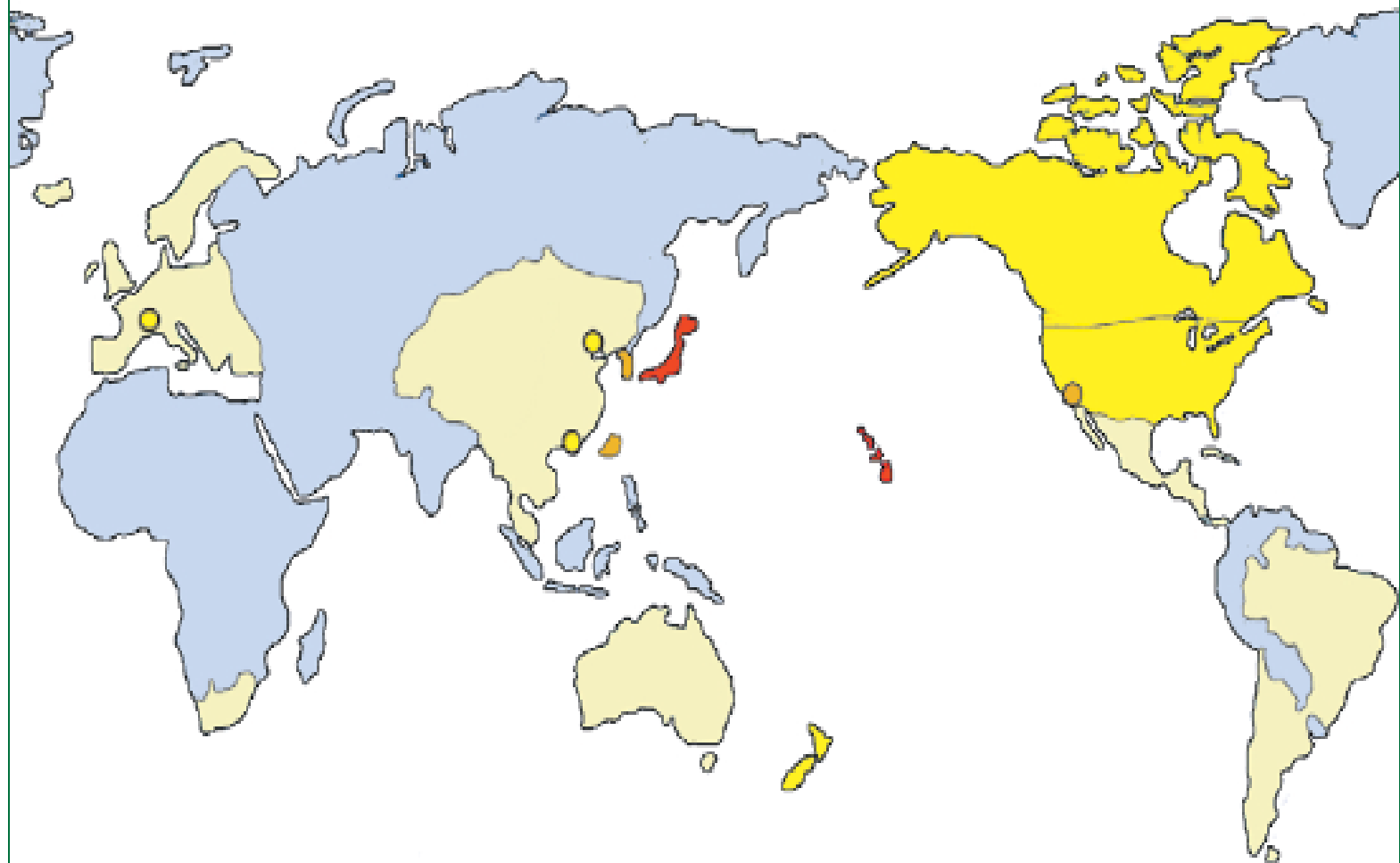
KAWASAKI DISEASE



KAWASAKI DISEASE

- First described in Japan in 1967 by Dr. Tomisaku Kawasaki
- Acute and self-limited vasculitis
- Infantile polyarteritis nodosa
- Mucocutaneous lymph node syndrome
- Surpassed acute rheumatic fever as the leading cause of acquired heart disease in children in the US

■ >100 ■ 50-100 ■ 10-50 ■ <10 ■ Unknown



1st Saudi Report

Annals of Tropical Paediatrics (1998) 18, 295–299

Kawasaki disease in a paediatric hospital in Riyadh

SAMEEH S. GHAZAL, MANSOUR ALHOWASI & MAGED M. EL SAMADY

Department of Paediatrics, Sulaimania Children's Hospital, Riyadh, Saudi Arabia

(Accepted 27 May 1998)

EPIDEMIOLOGY

- Positive family history in about 1%
- The rate in a sibling is 2.1% within 1 year after onset of the first case in a family
- The risk of occurrence in twins is 13%
- Incidence increased in children of parents who themselves had the illness in childhood

ETIOLOGY

■ Unknown

- Infectious, Genetic, Immune, Environmental?
- More common during the winter and early spring months in the US
- Occurs in epidemics with geographic wave like spread
- 76% of children are <5 years old with most cases in less than 2 years old

PATHOLOGY

- Generalized systemic vasculitis involving blood vessels throughout the body
- ^{self-limiting} Aneurysms may occur in other mid-size extraparenchymal muscular arteries such as the celiac, mesenteric, femoral, iliac, renal, axillary, and brachial arteries

manifestation : dilatation and aneurysm

aneurysm : one area dilated more than 150%

5 of surrounding vessels

DIAGNOSTIC CRITERIA

Table 1 Diagnostic criteria for Kawasaki disease.

Fever of at least 5 days duration plus four of the following features:

Bilateral conjunctival injection

Polymorphous exanthem

Changes in the lips and oral cavity

Cervical lymphadenopathy (> 1.5 cm)

Changes in the extremities:

Erythema of the hands and feet

Swelling of the hands and feet

Desquamation

Other diseases must be excluded

fever for 5 days

+ 4 out of this five

= diagnosis

conjunctiva

in lips

cervical lymphadenopathy

extremities

desquamation in hands and feet

strawberry tongue



DIAGNOSIS CRITERIA

- ≥ 5 days of fever and ≥ 4 of the 5 principal clinical features
- Typically, all of the clinical features are not present at a single point in time
- In the presence of 4 of 5 classic criteria, US and Japanese experts agree that only 4 days of fever are necessary before initiating treatment





A



C



E



B



D



F



LABORATORY FINDINGS

- Thrombocytosis appears in the second week the illness, peaks in the third week with a gradual return to normal by 4 to 8 wks
- Range from 500 000 to >1 million/mm³
- Thrombocytopenia is seen rarely in the acute stage and may be a sign of disseminated intravascular coagulation

LABORATORY FINDINGS

- Elevation of acute phase reactants (ESR and CRP) is universal
- Return to normal by 6 to 10 weeks
- The degree of elevation of ESR and CRP may show a discrepancy so both should be measured
- Elevation of ESR (but not of CRP) can be caused by IVIG therapy

CORONARIES

- 20% of untreated patients develop coronary aneurysm or ectasia
- 20% of aneurysms go on to develop stenosis
cardiac ischemia and infarction in YOUNG CHILD
- Ectasia or aneurysms regress in most patients within 1-2 years (AHA)
occur before age of 2 years
- Coronaries are ABNORMAL after aneurysms regression and even when aneurysms were never detected
and great majority within 5 years of life
- The long-term clinical implication is unknown

ECHOCARDIOGRAPHY

- Aneurysms are classified according to AHA statement as:

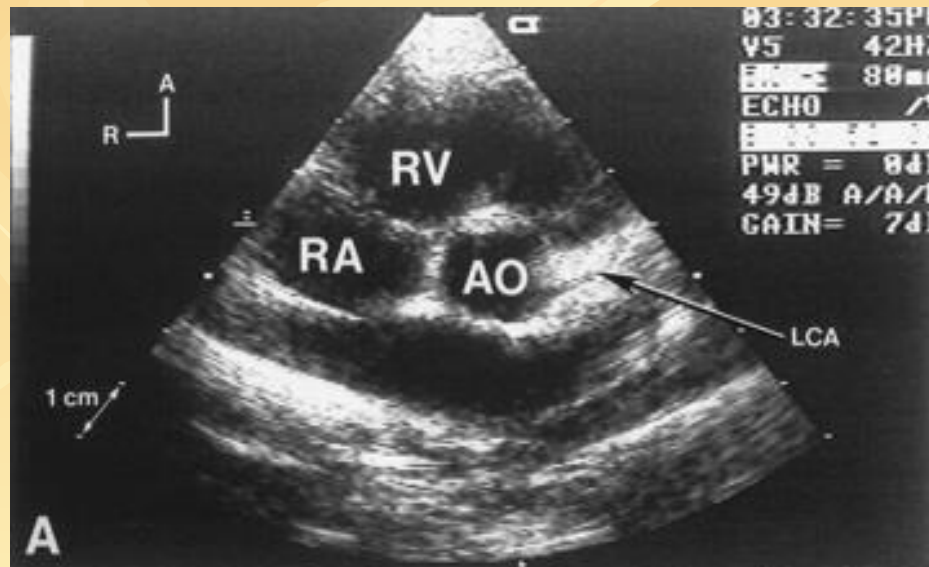
- small (<5-mm internal diameter),

- medium (5 to 8 mm internal diameter),

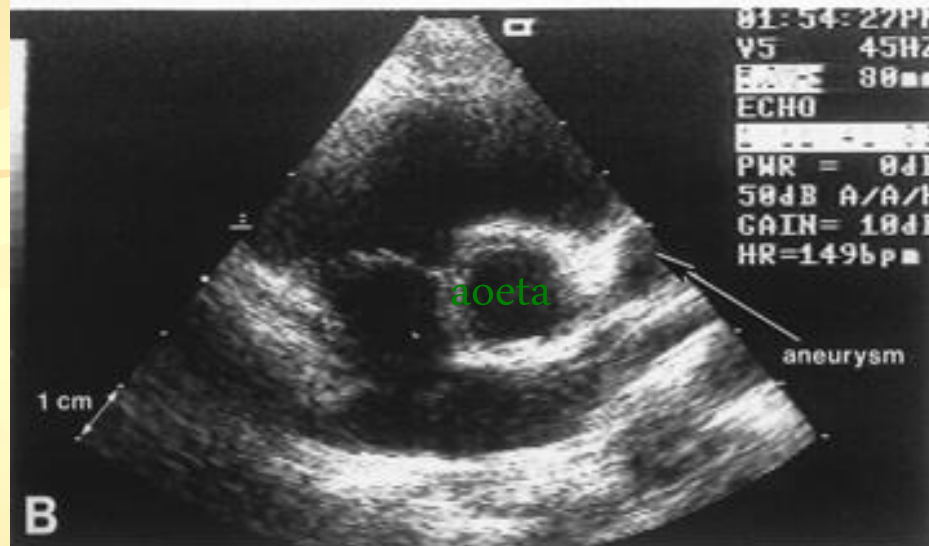
- or giant (>8-mm internal diameter).

not recovering spontinsly need cardiac surgery w\ poor prognosis





normal coronary artery



dilatation and aneurysm of coronary artery



angto // left and rt cornoary
almost same diameter of
aorta = sever dilation

MI: 1.6
S12
28 NOV 06
14:37:14
2/0/F/F3
KFMC

Pediatric
AL MOTAIRI
MOH'D
427019346

GAIN 54
COMP 59

6CM
120HZ

A +	DIST .443 cm
B X	DIST .404 cm
C -	DIST .278 cm

● Length = 0.28 cm
+ Length = 0.40 cm
■ Length = 0.44 cm

P T R
5 6 12
2.14
SEC

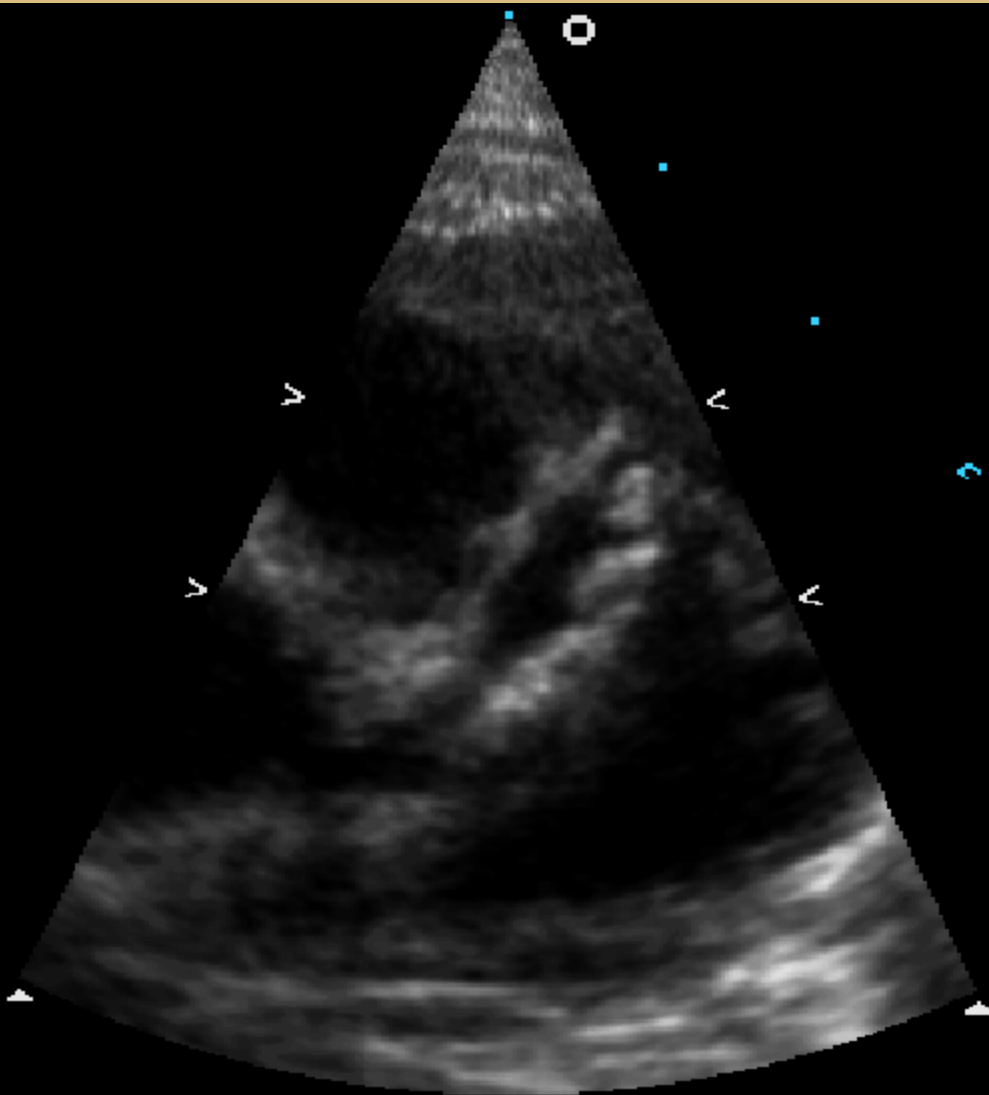
72
F 1 of 1

MI: 1.6
S8
20 SEP 06
13:03:08
2/2/E/F3
KFMC

Pediatric

GAIN 50
COMP 75

5CM
74HZ



T
P 3 R 8

TREATMENT

IVIG

most imp ttt .

- The efficacy of IVIG in reducing the prevalence of coronary artery abnormalities is well established
- Decreases the incidence of coronary aneurysms from 20% to 5% (if used before day 10) *best to give before day 8*
- Has generalized anti-inflammatory effect
- Patients should be treated with IVIG, 2 g/kg in a **single infusion over 10-12 hours** together with aspirin (AHA)

Durongpisitkul K, Gururaj VJ, Park JM, Martin CF. The prevention of coronary artery aneurysms in Kawasaki disease: a meta-analysis on the efficacy of intravenous gamma globulin and intravenous aspirin treatment. *Pediatrics*. 1997;100:1061-1064.

Coronary artery abnormalities in Kawasaki disease is highly dependent on gamma globulin dose but independent of salicylate dose. *J Pediatr*. 1997;131:888-893

TREATMENT

IVIG

- Give in the first 10 days and preferably within 7 days of illness
- Should be still administered to children presenting after the 10th day of illness if they have either
 - persistent fever without other explanation or
 - aneurysms and ongoing systemic inflammation, as manifested by elevated ESR or CRP
- Live vaccines deferred for 11 months

TREATMENT

ASPIRIN

Aspirin

- Additive anti-inflammatory effects to IVIG
- Does not appear to lower the frequency of the development of coronary abnormalities
- During the acute phase of illness, aspirin is administered at 80 to 100 mg/kg per day in 4 doses
- Many centers reduce the aspirin dose after the child has been afebrile for 48 to 72 hours. Other clinicians continue high-dose aspirin until day 14 of illness
- Low-dose aspirin (3–5 mg/kg per day) is maintained until the patient shows no evidence of coronary changes by 6 to 8 weeks after the onset of the illness *recomeded*
- For children who develop coronary abnormalities, aspirin may be continued indefinitely

Prognosis

- 50% of aneurysms regress within 5 years
- Mild dilatation (3–4 mm) regresses within 2 years
- 80% of those with moderate dilatation (4–8 mm) regress within 5 years.
- Giant aneurysms (>8 mm) are unlikely to resolve, and many progress to stenosis or complete obstruction within years of the initial diagnosis.